

# Two-dimensional echocardiography in infants with persistent truncus arteriosus\*

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**SUMMARY** Two-dimensional echocardiography was used to study a consecutive series of 13 infants with persistent truncus arteriosus. In all a single great artery with the long upward course characteristic of the aorta was shown. This great artery could be identified as a persistent truncus arteriosus by the recognition of a branch arising from its ascending part (10 out of 13) or of more than three semilunar valve cusps (5/13). These criteria allowed the correct diagnosis to be reached in 12 of the 13 infants.

The differentiation of persistent truncus arteriosus from other defects with a ventricular septal defect and overriding great artery can be difficult if not impossible with M-mode echocardiography. The differential diagnosis of this condition assumes particular importance in early infancy. We report the findings in a group of infants with persistent truncus arteriosus derived from our recent experience with two-dimensional echocardiography.

## Subjects and methods

Thirteen infants with persistent truncus arteriosus underwent echocardiographic studies before the diagnosis was established by angiocardiology. Eleven were newborn, aged from 4 hours to 9 days and the others, 5 and 11 weeks old; their weights ranged from 1.7 to 3.9 kg. In all but three the number of truncal valve cusps and the type of persistent truncus arteriosus were determined by operation or necropsy. In the remaining patients the type of persistent truncus arteriosus was determined by angiocardiology. The Table summarises the findings on angiocardiology, at surgery, at necropsy, and on echocardiography. The truncal anatomy was classified in accordance with the terminology of Collett and Edwards<sup>1</sup>; 10 were of type 1, two of type 2, and one of type 3. The truncal valve

was quadricuspid in four, and tricuspid in six of the cases where necropsy was performed.

During the study period another infant (27 days, 2.8 kg) was examined who had a ventricular septal defect, overriding aorta, pulmonary atresia with absent pulmonary arteries, and major aortopulmonary collateral arteries (type 4 truncus of Collett and Edwards). Though Van Praagh and Van Praagh<sup>2</sup> do

Table Comparison of echocardiographic appearances with findings subsequently obtained from angiocardiology, surgery, or necropsy

Patient		Angio/surgery/ necropsy		Echocardiography		
Age	Weight (kg)	Type	Cusps	Branches la sa	Cusps	LAd (mm)
1 d	2.5	3	4	—	2	4
11 wk	2.9	1	4	—	—	4
7 d	3.3	2	3	1	—	3
6 d	2.9	1	?	—	—	3
4 hr	3.3	1	3	1	1	3
6 d	3.4	2	3	—	1	3
9 d	2.8	1	3	—	1	3
7 d	1.7	1	3	1	1	3
5 wk	3.2	1	3	1	1	3
1 d	2.3	1	4	1	—	4
2 d	3.9	1	4	—	—	4
1 d	2.3	1	?	—	1	3
7 d	2.4	1	?	1	—	4

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Angio, angiocardiology; la, long axis view; sa, short axis view; LAd, left atrial dimension.

\* Necropsy information not available.

not consider this a true persistent truncus arteriosus the case is included because of possible difficulty in distinguishing it from persistent truncus arteriosus with M-mode echocardiography. The infant had monosomy 21 and cardiac catheterisation was not undertaken, the exact diagnosis being established by necropsy.

The echocardiographic equipment was a mechanical 60° sector scanner developed by the

Department of Clinical Physics and Bio-Engineering, Glasgow<sup>3,4</sup> or an ATL Mark III system.

A standard long axis view<sup>4,5</sup> showed the left ventricle and mitral valve, and was used to determine the relation of the upper ventricular septum to the great artery root. At end-systole the left atrial diameter was measured at semilunar valve level from the anterior margin of the posterior arterial wall to the posterior left atrial wall. This measurement was

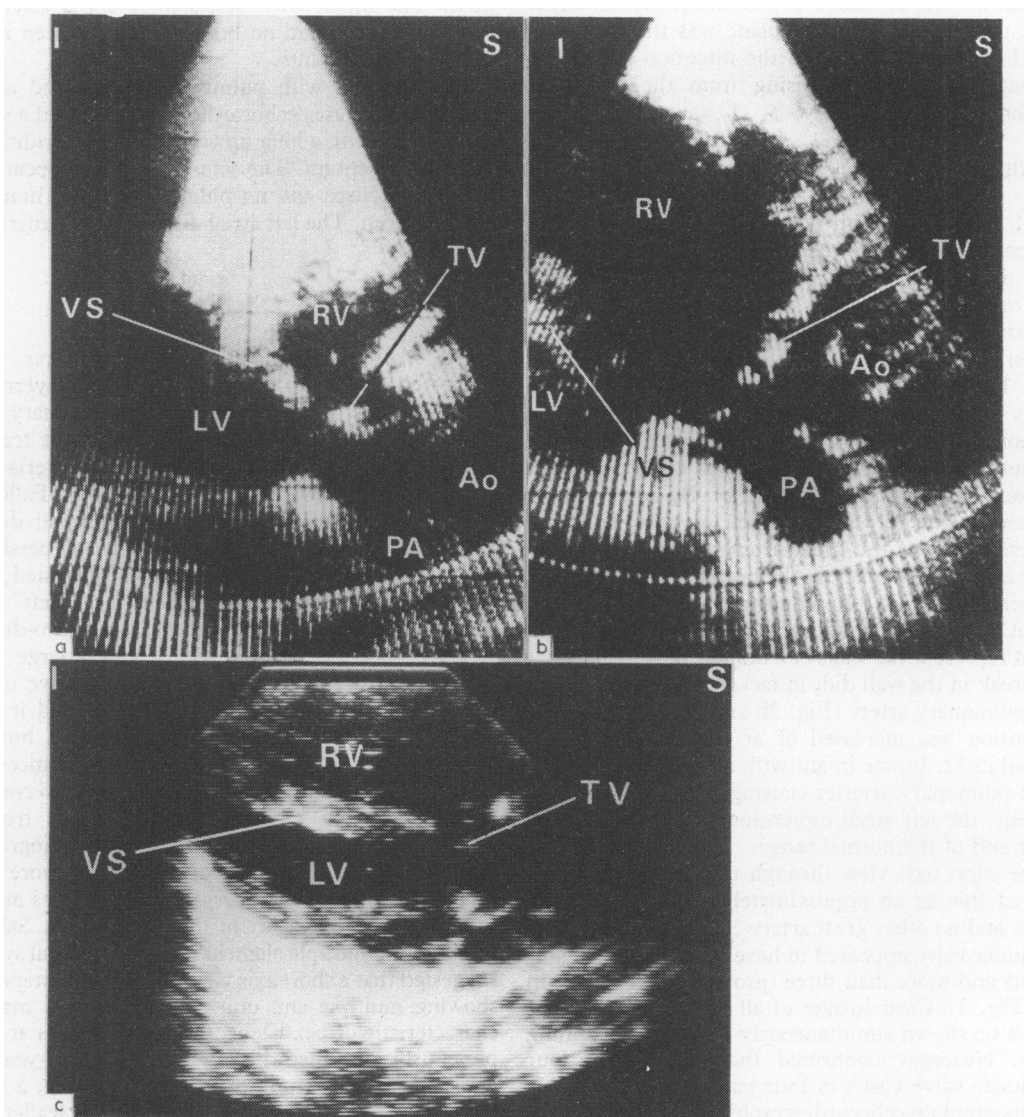


Fig. 1. Long axis views in diastole from infants with persistent truncus arteriosus. The truncus arises (a) almost equally from both ventricles, (b) mainly from the right ventricle, (c) mainly from the left ventricle. In (a) and (b) the division of the truncus arteriosus into aorta and pulmonary artery is clearly shown just above valve level. I, inferior; S, superior; Ao, aorta; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; TV, truncal valve leaflet; VS, ventricular septum.

considered to correspond to the left atrial dimension measured with M-mode echocardiography and for which normal values have been published.<sup>6</sup> In each case an average of six measurements was taken. A high long axis view<sup>7</sup> showed the upward course of the great artery and was tilted to the left and right as necessary in an attempt to show any branches arising from it. Short axis views at the base of the heart and through the great artery root were employed in an attempt to show another great artery, and then to provide as clear an image as possible of the semilunar valve cusps. The scanning plane was then tilted or edged upwards, again with the intention of demonstrating any branches arising from the ascending portion of the great artery.

## Results

In all 13 subjects with persistent truncus arteriosus echocardiography showed that the great artery overrode the ventricular septum; in diastole the vessel took origin almost equally from both ventricles in 10, almost entirely from the right ventricle in one, and almost entirely from the left ventricle in the other two (Fig. 1). The high long axis view showed the great artery to have a long upward course characteristic of the aorta (Fig. 2). In three patients its division into the aorta and pulmonary artery was easily shown in the long axis view (Fig. 1a and b). By adjusting the scanning plane to view as much as possible of the posterior wall of the great artery it was possible to show an apparent break in the posterior wall echo in a further six (Fig. 2a). In only three of these, however, was it possible to show clearly other echoes which might represent the walls of a branch and confirm that the break in the wall did, in fact, represent the origin of a pulmonary artery (Fig. 2b and c). The left atrial dimension was increased or at the upper range of normal in 12. In one infant with a type 3 truncus and small pulmonary arteries causing relative pulmonary stenosis, the left atrial dimension (8 mm) was at the lower end of the normal range.

The short axis view through the great artery root showed this as an approximately circular echo-free space, and no other great artery could be shown. The semilunar valve appeared to have three cusps in eight infants and more than three (probably four) cusps in five (Fig. 3). Clear images of all the cusps could not always be shown simultaneously in a single scanning plane. Necropsy confirmed the presence of four semilunar valve cusps in four patients in whom this was apparent on echocardiography, and of three cusps in six other subjects. By rotating or tilting the scanning plane upwards from the level of the semilunar valve a consistent break in the continuity of the circular wall echo was shown in seven infants. Since

failure to show a structure with ultrasound does not necessarily mean it is absent, care was taken to show that this gap in the wall represented the site of origin of the main (or a branch) pulmonary artery by the demonstration of two walls of the pulmonary artery in continuity with the great artery wall (Fig. 4). A single pulmonary artery arising from the posterior left aspect of the truncal root was seen in six infants (Fig. 4a). Two pulmonary artery branches arising quite separately from the left and right posterior walls were seen in the one infant with persistent truncus arteriosus type 3 (Fig. 4b) and no branch could be seen in the remaining six infants.

In the infant with pulmonary atresia and absent pulmonary arteries echocardiography showed a single great artery with a long upward course overriding the ventricular septum. The semilunar valve appeared to have three cusps and no pulmonary artery branches could be seen. The left atrial dimension was increased (16 mm).

## Discussion

With M-mode echocardiography it has been suggested that the demonstration of an overriding artery and failure to show the normal pulmonary valve echo may allow the diagnosis of persistent truncus arteriosus to be made.<sup>8</sup> Since these criteria are, however, satisfied in severe tetralogy of Fallot or pulmonary atresia with a ventricular septal defect, they are inadequate for the diagnosis of persistent truncus arteriosus unless, as has been suggested, they are found in association with a large left atrial dimension.<sup>9</sup> In these three conditions two-dimensional echocardiography demonstrates a large great artery with a long upward course suggestive of the aorta. If only one artery has been recorded it is of course conceivable that another is present but has merely escaped detection. In these circumstances it is therefore essential that an attempt is made to confirm or rule out the possibility of persistent truncus arteriosus. We have found two echocardiographic features of value in this: the presence of more than three cusps or of pulmonary artery branches arising from the ascending part of the great artery. Sahn *et al.*<sup>10</sup> using a multiple element two-dimensional system suggested that a short axis view of the great artery root showing multiple and unusual cusp echoes may be characteristic of persistent truncus arteriosus and we previously confirmed this appearance in a 15-year-old patient.<sup>11</sup> In this study of infants, however, a more detailed assessment of the semilunar valve leaflets has been possible. The number of valve cusps was correctly determined in all 10 cases of persistent truncus arteriosus where subsequent necropsy confirmation was available, including four with a quadri-

cuspid valve. The recognition of more than three semilunar cusps is diagnostic of persistent truncus arteriosus but in approximately 75% of cases where there is a bicuspid or tricuspid truncal valve<sup>2 12</sup> this diagnostic feature is absent. In these circumstances the diagnosis of persistent truncus arteriosus requires the demonstration of a branch (or branches) arising from the ascending part of the great artery. The identification of pulmonary artery branches in persis-

tent truncus arteriosus, however, poses some difficulties because, with two-dimensional echocardiography, a break in the great artery wall echo which is held to signify the origin of the pulmonary artery, or of a pulmonary artery branch, may be an artefact. Thus, before deciding that such a gap represents a pulmonary artery we consider it necessary to show the proximal part of the walls of the pulmonary artery and to show that these are in continuity with the adjacent

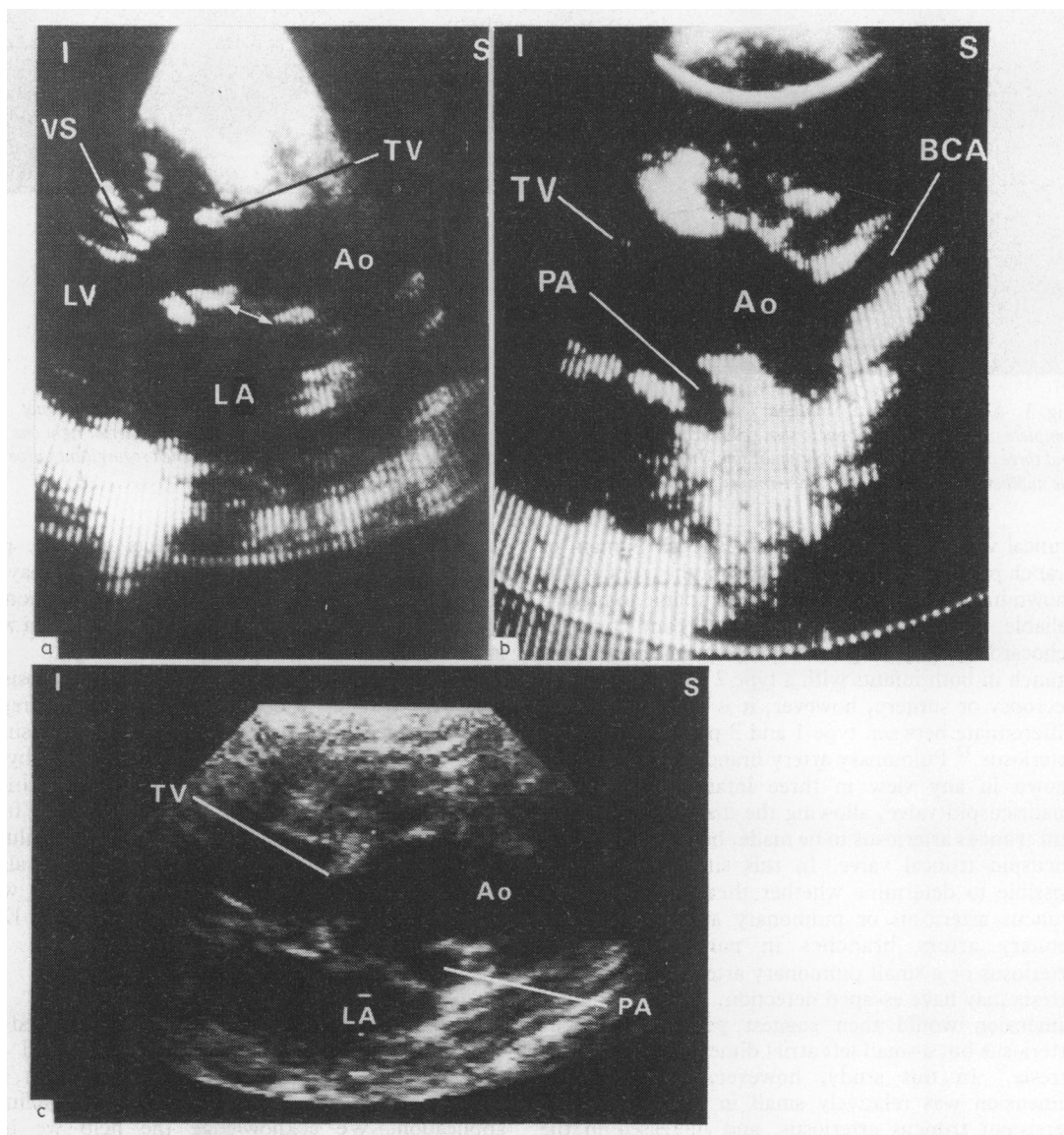


Fig. 2 Long axis views of ascending great artery. (a) A break in the posterior wall is apparent (indicated by the arrow) but a pulmonary artery cannot be demonstrated (b and c). The main pulmonary artery can be seen arising from the posterior wall of the great artery. BCA, brachiocephalic artery; LA, left atrium; VS, ventricular septum. Other abbreviations as before.

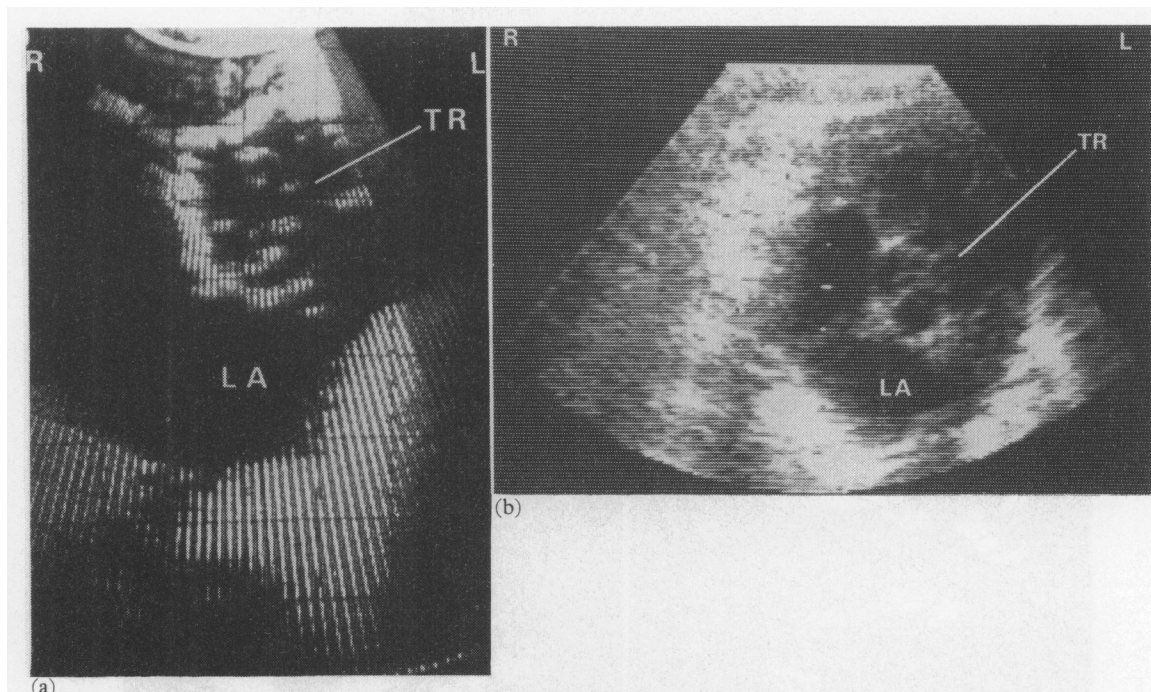


Fig. 3 Short axis views at semilunar level showing abnormal ( $\geq 4$ ) valve cusp echoes. The circular outline of the vessel root is not complete in these frames chosen to show cusp echoes. In (a) necropsy confirmed the presence of four cusps, a small posterior right one and three others of approximately equal size. In (b) retrograde truncal root angiocardiology showed a thickened regurgitant valve but the number of cusps could not be determined with certainty. R, right; L, left; LA, left atrium; TR, truncal root.

truncal wall (Fig. 2b and c, and Fig. 4). A main or branch pulmonary artery arising from the truncus was shown in 10 patients. We have been unable to make a reliable distinction between type 1 and type 2; echocardiography showed only one pulmonary artery branch in both infants with a type 2 truncus. Even at necropsy or surgery, however, it is often difficult to differentiate between type 1 and 2 persistent truncus arteriosus.<sup>12</sup> Pulmonary artery branches could not be shown in any view in three infants. Two had a quadricuspid valve, allowing the diagnosis of persistent truncus arteriosus to be made, but the third had a tricuspid truncal valve. In this situation it is not possible to determine whether there is a persistent truncus arteriosus or pulmonary atresia since pulmonary artery branches in persistent truncus arteriosus or a small pulmonary artery in pulmonary atresia may have escaped detection. A large left atrial dimension would then suggest persistent truncus arteriosus but a small left atrial dimension pulmonary atresia.<sup>9</sup> In this study, however, the left atrial dimension was relatively small in one infant with persistent truncus arteriosus, and increased in the infant with pulmonary atresia and major aortopulmonary collateral arteries of large size. Where there is no atrial septal defect or mitral valve disease

the left atrial size generally reflects only the pulmonary blood flow. Thus, the left atrium may be small in persistent truncus arteriosus with pulmonary branch stenosis or enlarged in pulmonary atresia with large aortopulmonary collateral flow.

We therefore consider that a definite diagnosis of persistent truncus arteriosus with echocardiography can be made only by the demonstration of a single great artery with a long upward course, and by its positive identification as a persistent truncus arteriosus by the recognition of a branch arising from its ascending part or of more than three semilunar valve cusps. Application of these criteria should allow the diagnosis to be reached in most infants with persistent truncus arteriosus, as was possible in 12 of the 13 infants in our series.

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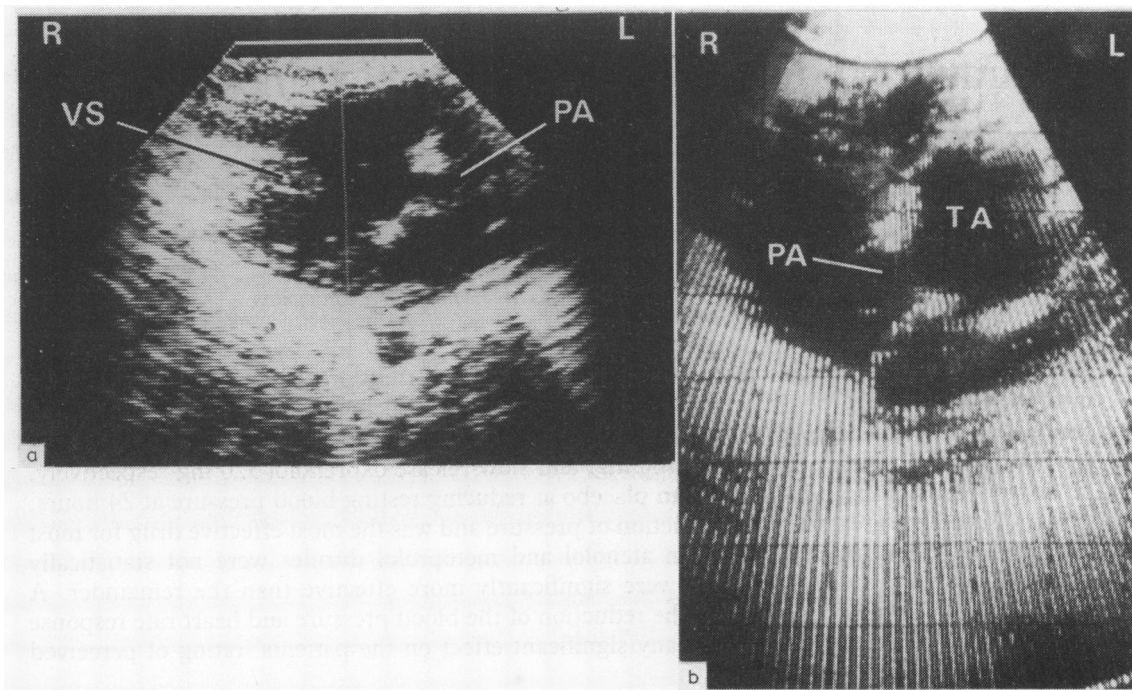


Fig. 4 Short axis views adjusted to show a branch arising from the truncus arteriosus. (a) Type 1 truncus; the main pulmonary artery is seen in a view rotated towards a long axis one and therefore showing only part of the truncal wall and the upper margin of the ventricular septum. (b) Type 3 truncus; the right pulmonary artery is shown in this view while the left one was shown by tilting the scanner to the left. TA, truncus arteriosus. Other abbreviations as before.

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